Compression therapy and liposuction of lower legs for bilateral hereditary primary lymphedema praecox

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In this report, we describe a case of bilateral non-syndromic hereditary lymphedema praecox of lower legs. The patient was diagnosed at age 16. Ten years later, he was unable to ambulate due to increased bilateral lower leg volume, continuous pain, and recurrent episodes of cellulitis. He was treated at our tertiary-care center with compression therapy and circumferential liposuction of lower legs, ankles, and dorsum of feet in order to remove hypertrophic fat deposits, facilitate conservative therapy, and decrease further risk of cellulitis. No complications were seen and compression therapy was continued. Fourteen month follow-up reveals no increase in leg volume over time, absence of pain, and no further episodes of cellulitis with complete ability to ambulate and return to normal activities. Even when it does not eliminate the underlying cause of primary lymphedema, combined therapy consisting of compression and liposuction is safe and is able to achieve control, at least on a short term, of clinically disabling conditions associated with advanced stages. (J Vasc Surg 2009;49:222-4.)

Advanced lower leg lymphedema presenting with adipose tissue hypertrophy may be unresponsive to manual lymphatic drainage-based complex decongestive therapy.[1,2] It progresses to dermato-lipo-fibroesclerotic conditions predisposing to recurrent episodes of cellulitis and, in rare cases, lymphangiosarcoma.[3-5] Therefore, excisional surgery through long incisions has been advocated, but it has been either inadequate or required multiple procedures associated with scarring, wound complications, dysesthesias, and a cumbersome postoperative course.[6,7] These situations have been overcome in arm secondary lymphedema by the use of liposuction through small incisions.[8] Few reports on its use in patients with lower leg lymphedema have been published and there is no data specifically addressing outcomes regarding different forms of primary lymphedema.

The purpose of this report is to evaluate short-term outcomes of compression therapy and liposuction on a patient with bilateral lower leg primary lymphedema praecox.

CASE REPORT

A 26-year old man with non-syndromic lymphedema of both lower legs since age 16 had been managed for 10 years with manual lymphatic drainage and 20 mmHg ace wraps as his best community treatment available. During the last five years, he had developed 10 episodes of cellulitis requiring antibiotics and was in continous pain not alleviated by medications. Previously performed lymphoscintigraphy showed groin uptake more than two hours after injection of feet. He was brought to our tertiary-care center where he was found to have stage III bilateral lower leg lymphedema with non-pitting elephantine trophic changes in the skin, acanthosis, large fat deposits, and crippling invalidism.[9] Lower leg circumferences at different levels were measured and found to be larger at mid-calves where right and left legs were 76 cm and 68 cm, respectively. Volumes were determined following the disc model method.[10,11] Right and left volumes were 18.4 liters and 14.7 liters, respectively. Computed tomography (CT) scan of chest, abdomen, pelvis, and groin was normal. Bilateral leg elevation and 40 mmHg compression bandages were initiated. One week later, leg circumference at mid-calves was 58 cm on the right side and 53 cm on the left side (Figs 1 and 2, left), and volumes were 10.7 L and 8.9 L, respectively. At this point in time, liposuction was performed in order to remove hypertrophic adipose deposits, facilitate conservative therapy, and reduce further risk of cellulitis. The procedure began by infiltrating 1000 mL of Ringer's lactate solution with 250 mg of lidocaine and 1 mg of epinephrine 1:1000 in the subcutaneous space of both lower legs, ankles, and dorsum of feet. After allowing for proper vasoconstriction, circumferential suctioning of all fat deposits was performed through three 3-mm incisions on each leg until skin and subcutaneous tissue consistencies were soft and smooth. Total aspirate from the right side was 1650 mL, consisting of 1000 mL of fat, 600 mL of injectate fluid, and 50 mL of blood. Total aspirate from the left side was 1250 mL and consisted of 600 mL of fat, 600 mL of injectate fluid, and 50 mL of blood. Suction drains were placed and limbs were wrapped up with 40 mmHg compression bandages. Patient was kept on bilateral leg elevation, allowing for showering and limited ambulation.

There were no complications during the surgical procedure and during the hospitalization. Patient was discharged two days after surgery. Drains and prophylactic antibiotics were discontin-

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Fig 1. Anterior view of lower legs 1 week after therapy with 40 mm Hg compression bandages and elevation (left) and 12 months after circumferential liposuction and continuous compression therapy (right).

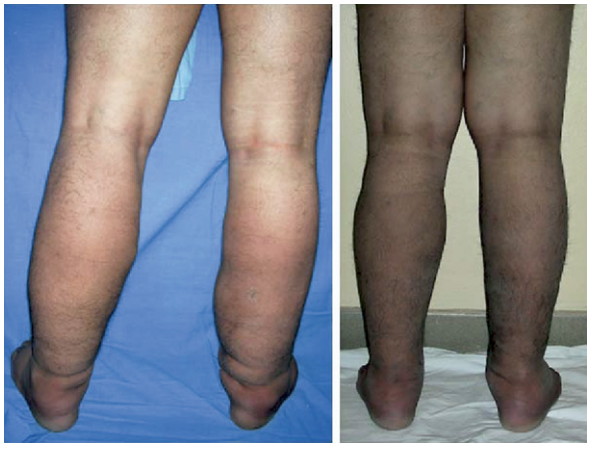

Fig 2. Posterior view of lower legs 1-week after therapy with 40 mm Hg compression bandages and elevation (left) and 12 months after circumferential liposuction and continuous compression therapy (right).

ued two weeks afterward. There have been no complications during his follow-up, which is 14 months now. Postoperative right and left lower leg circumferences are 45 cm and 44 cm, respectively and volumes are 6.4 L and 6.1 L, respectively (Figs 1 and 2, right). Patient is still on manual lymphatic drainage and wearing 40 mm Hg compression bandages. There has been no increase in lower leg volume and he is totally able to ambulate. Skin care with topical 10% urea ointment has maintained skin softness. Cellulitis has not occurred and antibiotics have not been required so far.

DISCUSSION

Multiple therapies have been designed to improve clinical conditions in primary lower leg lymphedema.[12] Manual lymphatic drainage-based complex decongestive therapy has been proven to be the single most effective method to control lymphedema and prevent its complications.[13] It is especially effective during early clinical stages and remains a critical part of the multimodal therapy required for advanced cases.[14] Creation of lymphovenous anastomosis has been of benefit only in patients with secondary lymphedema who have dilated lymphatic vessels and in the early stages while there are salvageable lymph vessels available for reconstruction.[15] Debulking methods reduce the amount of hypertrophic fat present in lymphedematous limbs and have been very helpful in facilitating conservative therapy, improving function, and preventing further episodes of cellulitis.[16] Fat removal has typically been accomplished through long incisions with its associated morbidity. One of these methods, the Charles procedure, consisted of skin, fat, and fascia removal with resurfacing with a skin graft. This procedure often caused worsening conditions, ultimately requiring limb amputations and therefore is no longer performed these days.[7] Staged excisions of skin and fat have been very helpful in improving symptoms but they require more than one procedure and leave patients with the possibility of wound complications, dysesthesias, scarring, and a cumbersome postoperative course due to intensive care of surgical wounds.[6] Liposuction removes large volumes of hypertrophic fat deposits without requiring long skin incisions. It has proven to be very useful in secondary lymphedema of the arms, particularly in advanced stages when lymph vessels are almost completely non-functioning.[8,17] Concerns have risen about the possible trauma that liposuctioning might cause to fragile lymph vessels and subsequent worsening of lymphedema. To date, there has been no clinical, anatomical, or experimental evidence of such an effect. One explanation to this is that in advanced stages, lymph vessels are already almost completely non-functioning and the benefits of volume reduction are clinically more evident than the possible worsening of the remaining, if any, vessel function. To date, there are three reports in the literature on the use of liposuction as the unique surgical modality in lower leg lymphedema. The first two of them involved five patients each, with unilateral secondary lymphedema.[18,19] The third one included one patient with bilateral primary lymphedema tarda.[20] The degree of benefit has not been uniform. It appears that maintenance of compression therapy is of paramount importance for two reasons: first, to avoid subsequent accumulation of fluid; and second, to favor retraction of expanded and hypertrophic skin.

In this report, a patient with primary lower leg lymphedema praecox was managed bilaterally with compression therapy and circumferential liposuction. This was accomplished in order to remove hypertrophic adipose deposits, facilitate conservative therapy, and decrease further episodes of cellulitis. Blood loss was minimal by infiltrating subcutaneous space with tumescent solution. Wound care was simple and rehabilitation was prompt. Manual massaging and use of multilayered compression bandages is mandatory to aid in skin retraction and avoid subsequent fluid accumulation. In this case, liposuction

allowed for conservative treatment to be more effective, decreased volume and therefore pain, reduced further risk of cellulitis, improved skin quality, and facilitated skin care. Short-term follow-up has not shown complications or recurrence of lymphedema-related conditions. We conclude that, even when circumferential liposuction does not eliminate underlying anomalies seen in primary lymphedema, it is safe and effective in facilitating conservative therapy to control disabling conditions associated with advanced stages.

AUTHOR CONTRIBUTIONS

Conception and design: AE-M, CAH, MI
Analysis and interpretation: AE-M, CAH, LA, MI
Data collection: AE-M, CAH, LA, MI
Writing the article: AE-M
Critical revision of the article: AE-M, CAH, LA, MI
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